Case Report

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Primary solitary fibrous tumour in the pulmonary artery: a case report

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Abstract

This report presents a very rare case of fibrous tumour in the pulmonary artery. A 32-year-old male patient presented at the First Hospital of Jilin University with the chief complaint of chest pain accompanied by cough and haemoptysis. Laboratory test results showed a D-dimer level of 60 ng/ml, and protein S activity of 51.0%. Echocardiography and enhanced computed tomography (CT) of the lung revealed a thrombus-like mass in the main pulmonary artery trunk, and the left pulmonary artery and its branches. The patient was initially diagnosed with pulmonary embolism, but after performing arterial endarterectomy, the presence of a tumour in the corresponding areas was confirmed. Histopathology and immunohistochemistry indicated the presence of a malignant solitary fibrous tumour. Chemotherapeutic agents were administered following surgery. The patient was prescribed ongoing oral Apatinib (250 mg, once daily) as a tumour-targeting therapy. The patient refused permission for postoperative CT. The patient was still alive at the 2-year follow-up.

Keywords

Solitary fibrous tumour, right heart catheterization, endarterectomy

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Introduction

Primary solitary fibrous tumour in the pulmonary artery is a rare condition with non-specific clinical manifestations.[1–3] First described by Klemperer and Rabin in 1931,[4] solitary fibrous tumours most

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commonly originate from the pleura, however, extrapleural anatomic locations have also been reported.[5] Notably, a solitary fibrous tumour in the pulmonary artery might mimic pulmonary embolism.[1,6,7] Immunohistochemical staining of solitary fibrous tumour tissue reveals positive signals for vimentin, CD34 antigen, CD99 antigen, and signal transducer and activator of transcription 6 (STAT6).[8–10]

Case report

A 32-year-old male was admitted to the Emergency Department of the First Hospital of Jilin University in February 2017 with the chief complaint of chest pain for the previous 8 months, accompanied by cough and haemoptysis for the previous 2 weeks. The patient's blood pressure was 132/80 mmHg, the pulse rate was 84 beats per min, and the physical examination did not reveal significant findings. Laboratory test results showed a D-dimer level of 60 ng/ml (reference range, 0–232 ng/ml), a high-sensitivity C-reactive protein level of 12.2 mg/l (reference range, 0–3.5 mg/l) and level of plasma protein S activity of 51.0% (reference range, 60–130%). Other laboratory results were within normal ranges. Echocardiography, performed using a Philips iE33 colour Doppler ultrasound system (Philips Healthcare, Andover, MA, USA), revealed that the left atrium was 33 mm, the right atrium was enlarged (40 mm), and the main pulmonary artery was widened (30 mm). A hyperechogenic mass was detected in the main pulmonary artery bifurcation and the origin of the left pulmonary artery, the size of which was measured to be 40×16 mm. Enhanced computed tomography (CT) of the lung, performed using a Discovery CT750 HD CT scanner (GE Healthcare, Chicago, IL, USA), revealed a mass, shown as a filling defect, in the pulmonary arterial trunk, the left main pulmonary artery and its distal branches, with a CT density value of 37 Hounsfield Units (non-enhanced; Figure 1). Venous compression ultrasonography (Philips iU22 colour Doppler ultrasound system; Philips Healthcare) of the bilateral lower limbs showed no abnormal findings, and the initial diagnosis was pulmonary thromboembolism. After consultation with a multidisciplinary team regarding pulmonary embolism, a pulmonary arterial tumour was suspected. Positron emission tomography (PET)-CT, performed using a Siemens Biograph 16 HR PET/CT system (Siemens Healthcare GmbH, Erlangen,

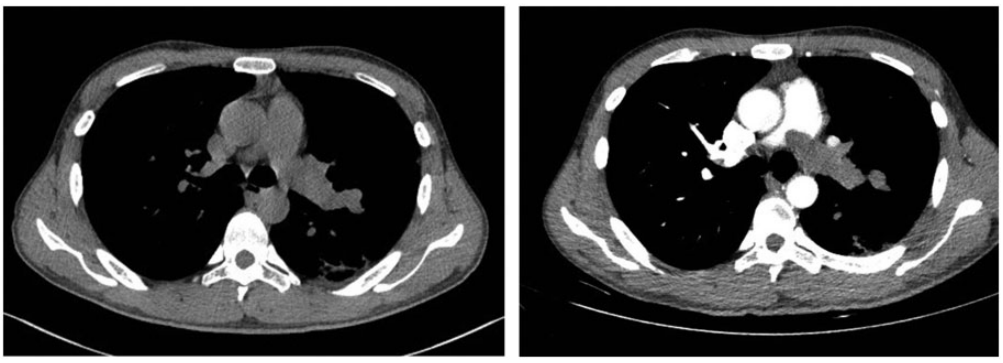

Figure 1. Computed tomography (CT) image of the lung (left) and enhanced CT image of the lung (right) revealing a mass, shown as a filling defect, in the pulmonary arterial trunk, left main pulmonary artery and its distal branches. In the non-enhanced CT, the mass had a CT density value 37 Hounsfield Units.

Germany), showed a hypermetabolic nodal mass in the main pulmonary artery trunk and the left pulmonary artery. The size of the biggest node measured was 1.5 cm × 1.4 cm, with a maximum standard uptake value (SUVmax) of 5.4 (Figure 2). Pulmonary arterial endarterectomy was then performed and revealed a tumour that completely occluded the left pulmonary artery, extending to the main pulmonary artery and distal branches of left main pulmonary artery. Histopathology of the tumour tissue revealed a malignant spindle-shaped tumour. Subsequent immunohistochemistry showed that the tumour tissue was positive for vimentin, CD34 antigen and CD99 antigen. The final diagnosis was primary malignant solitary fibrous tumour in the pulmonary artery. Following surgery, six 21-day treatment cycles of 2.0 g ifosfamide, i.v., day 1–3, and 100 mg epirubicin civ 96 h, were administered. The response was evaluated as stable disease. The patient was then prescribed 250 mg apatinib, oral, once daily, ongoing, as a tumour targeted therapy. The patient refused to give permission for postoperative CT. At the 2-year follow-up the patient was still alive.

The requirement for ethics approval to publish this case report was waived by the Institutional Review Board of the First Hospital of Jilin University. Written informed consent to publish the case was provided by the patient.

Discussion

Pulmonary artery sarcoma is a rare condition that is usually misdiagnosed as a pulmonary thromboembolism, and is even managed with anticoagulation or thrombolytic therapy.[1,6,7] The pathological types of primary pulmonary arterial tumour include leiomyosarcoma, malignant fibrous histiocytoma, fibrosarcoma and solitary fibrous tumour.[11] The solitary fibrous tumour is a rare mesenchymal neoplasm, and although it most commonly originates from the pleura, there have been reports of extrapleural anatomic locations.[5,12] In numerous cases, solitary fibrous tumour in the pulmonary artery has been reported to mimic pulmonary embolism.[1,6,7] In the present case, the primary complaints were chest pain and haemoptysis, and echocardiography and lung enhanced CT revealed a

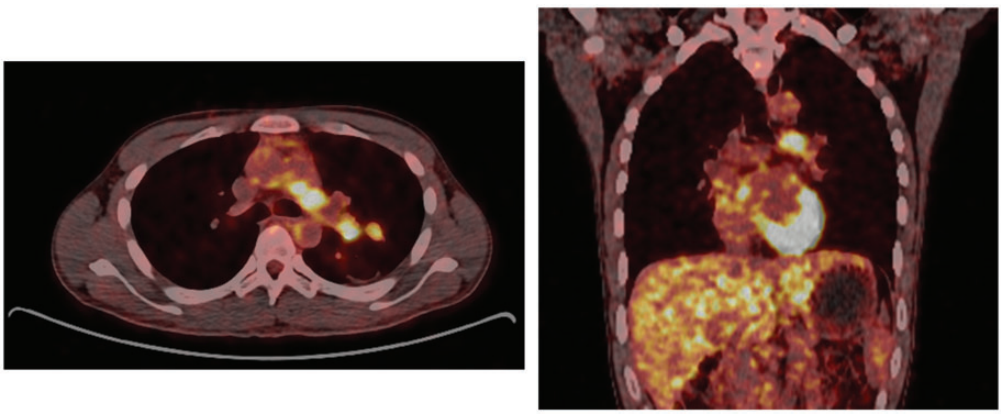

Figure 2. Positron emission tomography-computed tomography images (cross-section [left] and coronal section [right]) showing a hypermetabolic nodal mass in the main pulmonary artery trunk and the left pulmonary artery. The biggest node measured was 1.5 cm × 1.4 cm, with a maximum standard uptake value (SUVmax) of 5.4.

thrombus-like mass in the main and left pulmonary artery. These findings conformed to the clinical features of pulmonary embolism; however, D-dimer was within the normal range, and no deep venous thrombus was found. In addition, the echocardiogram did not show an increase in pulmonary arterial pressure. Features within the CT images of the pulmonary artery tumour showed some differences with an arterial thrombus. For example. the chronic thrombus is generally ring shaped or crescent type, and makes an obtuse angle with the blood vessel wall, while the tumour mostly manifests as a non-uniform filling defect in the pulmonary artery. The PET-CT images in the present case revealed a nodal mass with high radioactive uptake in the pulmonary artery, which is commonly associated with malignant neoplasm, and is consistent with other reports.[13,14] Confirmation of the diagnosis relies on the histopathological and immunohistochemical findings. Solitary fibrous tumour tissue shows spindle-shaped cells in a variable collagen stroma, which is normally positive for vimentin, CD34, CD99, and STAT6.[8] The prognosis of pulmonary artery sarcoma is generally poor, with a mean survival duration of about 1 to 2 months without therapy, extending to 8–36 months after surgical treatment.[15] In the present case, after surgical treatment and chemotherapy, and at two years of follow-up, the patient was still alive.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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